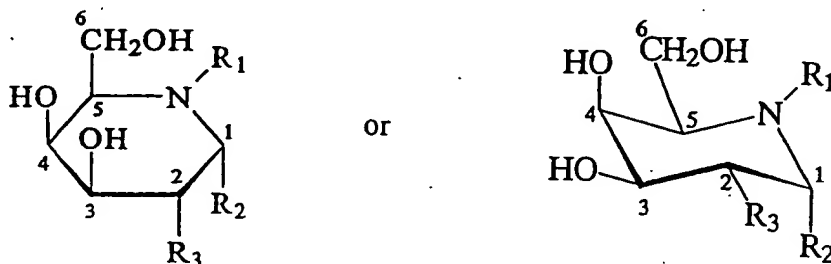


IN THE CLAIMS

Please cancel claims 1-9. Please add new claims 10-48 as follows:

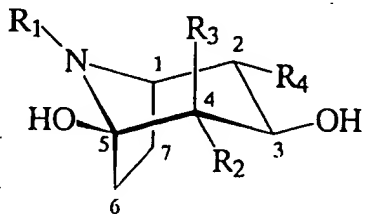
1-9. Canceled

10. (new) A method of treating Fabry disease comprising administering to an individual in need thereof an effective amount of a compound of the formula:



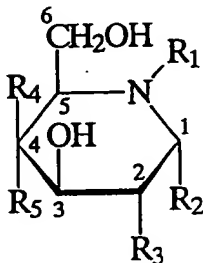
wherein R₁ represents H, CH₃, or CH₂CH₃; and
R₂ and R₃ independently represent H, OH, a simple sugar, a 1-3 carbon alkyl, alkoxyl, or hydroxyalkyl group.

11. (new) A method of treating Fabry disease comprising administering to an individual in need thereof an effective amount of a compound of the formula:



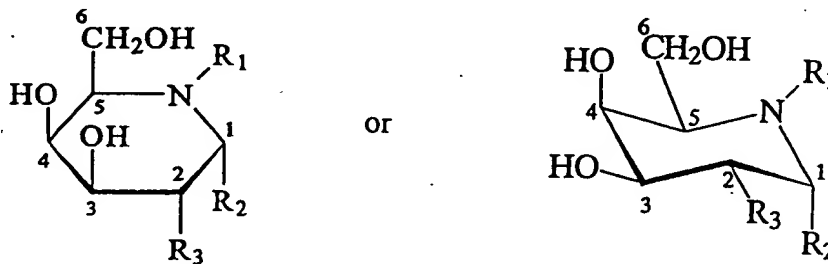
wherein for calystegine A₃: R₁ = H, R₂ = OH, R₃ = H, R₄ = H;
for calystegine B₂: R₁ = H, R₂ = OH, R₃ = H, R₄ = OH;
for calystegine B₃: R₁ = H, R₂ = H, R₃ = OH, R₄ = OH; and
for N-methyl-calystegine: R₁ = CH₃, R₂ = OH, R₃ = H, R₄ = H.

12. (new) A method of treating Fabry disease comprising administering to an individual in need thereof an effective amount of a compound of the formula:



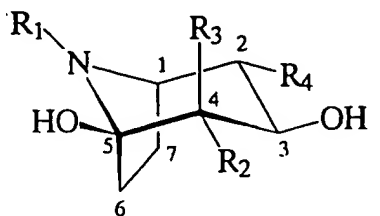
wherein R_1 represents H, CH_3 , or CH_2CH_3 ;
 R_2 and R_3 independently represent H, OH, a 1-6 carbon alkyl, hydroxyalkyl, alkoxy, or a simple sugar; and
 R_4 and R_5 independently represent H or OH.

13. (new) A method of enhancing the activity of lysosomal α -galactosidase A in a mammalian comprising administering an effective amount of a compound of the formula:



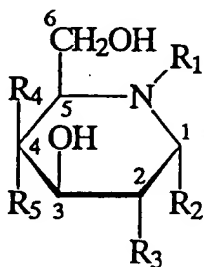
wherein R_1 represents H, CH_3 , or CH_2CH_3 ; and
 R_2 and R_3 independently represent H, OH, a simple sugar, a 1-3 carbon alkyl, alkoxy, or hydroxyalkyl group.

14. (new) A method of enhancing the activity of lysosomal α -galactosidase A in a mammalian comprising administering an effective amount of a compound of the formula:



wherein
 for calystegine A₃: R₁ = H, R₂ = OH, R₃ = H, R₄ = H;
 for calystegine B₂: R₁ = H, R₂ = OH, R₃ = H, R₄ = OH;
 for calystegine B₃: R₁ = H, R₂ = H, R₃ = OH, R₄ = OH; and
 for N-methyl-calystegine: R₁ = CH₃, R₂ = OH, R₃ = H, R₄ = H.

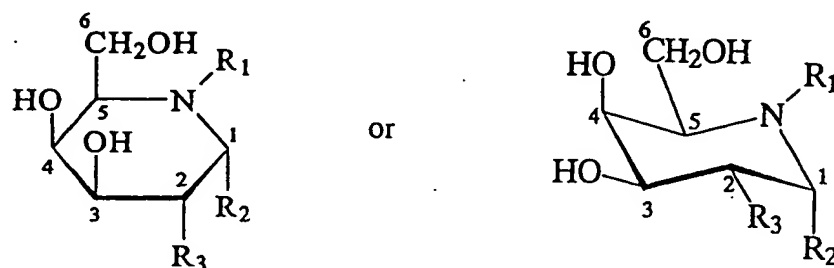
15. (new) A method of enhancing the activity of lysosomal α -galactosidase A in a mammalian comprising administering an effective amount of a compound of the formula:



wherein
 R₁ represents H, CH₃, or CH₂CH₃;
 R₂ and R₃ independently represent H, OH, a 1-6 carbon alkyl, hydroxyalkyl, alkoxy, or a simple sugar; and
 R₄ and R₅ independently represent H or OH.

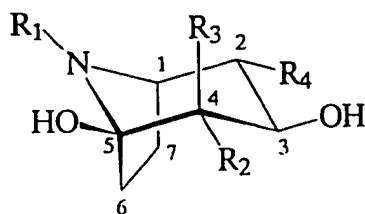
16. (new) A method of stabilizing lysosomal α -galactosidase A in mammalian cells comprising administering an effective amount of a compound of formula:

16. (new) A method of stabilizing lysosomal α -galactosidase A in mammalian cells comprising administering an effective amount of a compound of formula:



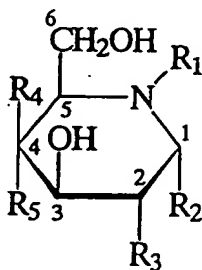
wherein R_1 represents H, CH_3 , or CH_2CH_3 ; and
 R_2 and R_3 independently represent H, OH, a simple sugar, a 1-3 carbon alkyl, alkoxy, or hydroxyalkyl group.

17. (new) A method of stabilizing lysosomal α -galactosidase A in mammalian cells comprising administering an effective amount of a compound of formula:



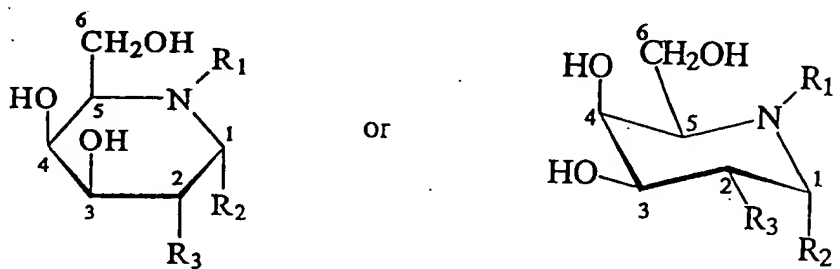
wherein for calystegine A₃: $R_1 = H$, $R_2 = OH$, $R_3 = H$, $R_4 = H$;
 for calystegine B₂: $R_1 = H$, $R_2 = OH$, $R_3 = H$, $R_4 = OH$;
 for calystegine B₃: $R_1 = H$, $R_2 = H$, $R_3 = OH$, $R_4 = OH$; and
 for N-methyl-calystegine: $R_1 = CH_3$, $R_2 = OH$, $R_3 = H$, $R_4 = H$.

18. (new) A method of stabilizing lysosomal α -galactosidase A in mammalian cells comprising administering an effective amount of a compound of formula:



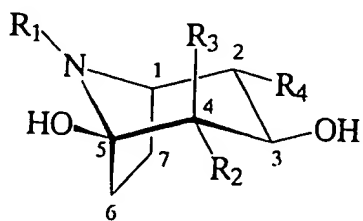
wherein R_1 represents H, CH_3 , or CH_2CH_3 ;
 R_2 and R_3 independently represent H, OH, a 1-6 carbon alkyl, hydroxyalkyl, alkoxy, or a simple sugar; and
 R_4 and R_5 independently represent H or OH.

19. (new) A method of preventing the degradation of lysosomal α -galactosidase A in a mammalian comprising administering an effective amount of a compound of the formula:



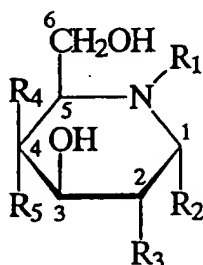
wherein R_1 represents H, CH_3 , or CH_2CH_3 ; and
 R_2 and R_3 independently represent H, OH, a simple sugar, a 1-3 carbon alkyl, alkoxy, or hydroxyalkyl group.

20. (new) A method of preventing the degradation of lysosomal α -galactosidase A in a mammalian comprising administering an effective amount of a compound of the formula:

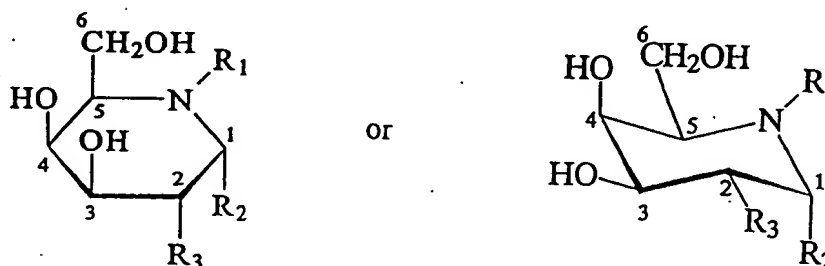


wherein for calystegine A₃: $R_1 = H$, $R_2 = OH$, $R_3 = H$, $R_4 = H$;
for calystegine B₂: $R_1 = H$, $R_2 = OH$, $R_3 = H$, $R_4 = OH$;
for calystegine B₃: $R_1 = H$, $R_2 = H$, $R_3 = OH$, $R_4 = OH$; and

21. (new) A method of preventing the degradation of lysosomal α -galactosidase A in a mammalian comprising administering an effective amount of a compound of the formula:

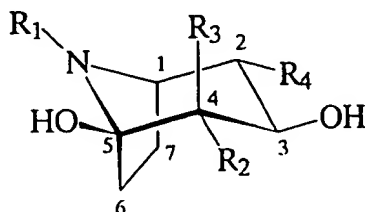


22. (new) A method of preventing deposition of neutral glycosphingolipids in vascular endothelial cells in an individual in need thereof, comprising administering an effective amount of a compound of formula:



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23. (new) A method of preventing deposition of neutral glycosphingolipids in vascular endothelial cells in an individual in need thereof, comprising administering an effective amount of a calystegine compound of formula:



wherein

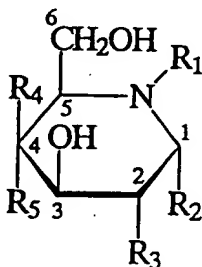
for calystegine A₃: R₁= H, R₂=OH, R₃=H, R₄=H;

for calystegine B₂: R₁= H, R₂=OH, R₃=H, R₄=OH;

for calystegine B₃; R₁= H, R₂=H, R₃=OH, R₄=OH; and

for N-methyl-calystegine: R₁= CH₃, R₂=OH, R₃=H, R₄=H.

24. (new) A method of preventing deposition of neutral glycosphingolipids in vascular endothelial cells in an individual in need thereof, comprising administering an effective amount of a compound of formula:



wherein

R₁ represents H, CH₃, or CH₂CH₃;

R₂ and R₃ independently represent H, OH, a 1-6 carbon alkyl, hydroxyalkyl, alkoxy, or a simple sugar; and

R₄ and R₅ independently represent H or OH.

25. (new) The method of claim 24, wherein the compound is selected from the group consisting of 1-deoxynojirimycin, 1-deoxygalactonojirimycin, α -homonojirimycin, 3,4-diepi- α -homonojirimycin, and 4-*epi*-fagomine.

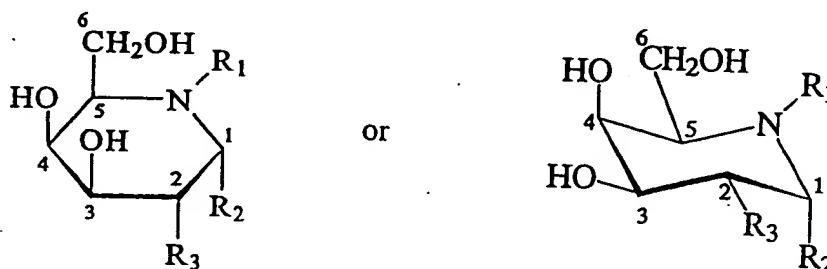
26. (new) The method of claim 25 wherein the compound is 1-deoxygalactonojirimycin.

27. (new) The method of claim 22, wherein the glycosphingolipids are predominantly ceramide trihexoside.

28. (new) The method of claim 23, wherein the glycosphingolipids are predominantly ceramide trihexoside.

29. (new) The method of claim 24, wherein the glycosphingolipids are predominantly ceramide trihexoside.

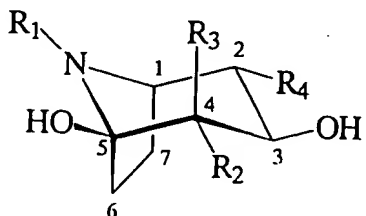
30. (new) A method of preventing renal failure associated with deposition of neutral glycosphingolipids in vascular endothelial cells in an individual in need thereof, comprising administering an effective amount of a compound comprising administering an effective amount of a compound of formula:



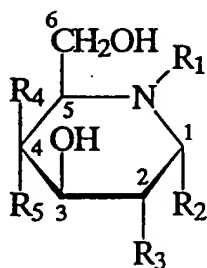
wherein R₁ represents H, CH₃, or CH₂CH₃; and

R₂ and R₃ independently represent H, OH, a simple sugar, a 1-3 carbon alkyl, alkoxyl, or hydroxy-alkyl group.

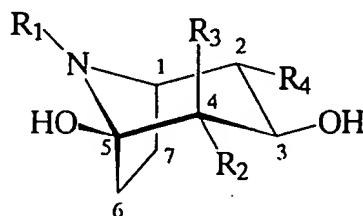
The diagram shows a bicyclic molecule, specifically a 1,2,3,4,5,6,7-heptamethylene-2,3-diol derivative. The structure is numbered 1 through 7. Substituents include R_1 on the nitrogen atom, R_2 on carbon 4, R_3 on carbon 2, and R_4 on carbon 3. Hydroxyl groups (OH) are attached to carbons 3 and 5. The stereochemistry is indicated by wedge and dash bonds: the OH group on carbon 5 is on a wedge, while the OH group on carbon 3 is on a dash. The nitrogen atom is part of a five-membered ring fused to a six-membered ring.



Chemical structure of a substituted piperidine derivative. The piperidine ring is numbered 1 to 6. Substituents include R_1 at N, R_2 at C1, R_3 at C2, R_4 at C4, R_5 at C5, and a CH_2OH group at C6. An OH group is attached to C4.



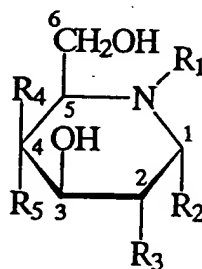
39. (new) A method of preventing premature myocardial infarctions and strokes associated with deposition of neutral glycosphingolipids in vascular endothelial cells in an individual in need thereof, comprising administering an effective amount of a compound of formula:



wherein

- for calystegine A₃: R₁ = H, R₂ = OH, R₃ = H, R₄ = H;
- for calystegine B₂: R₁ = H, R₂ = OH, R₃ = H, R₄ = OH;
- for calystegine B₃: R₁ = H, R₂ = H, R₃ = OH, R₄ = OH; and
- for N-methyl-calystegine: R₁ = CH₃, R₂ = OH, R₃ = H, R₄ = H.

40. (new) A method of preventing premature myocardial infarctions and strokes associated with deposition of neutral glycosphingolipids in vascular endothelial cells in an individual in need thereof, comprising administering an effective amount of a compound of formula:



wherein

- R₁ represents H, CH₃, or CH₂CH₃;
- R₂ and R₃ independently represent H, OH, a 1-6 carbon alkyl, hydroxyalkyl, alkoxy, or a simple sugar; and
- R₄ and R₅ independently represent H or OH.

41. (new) The method of claim 40, wherein the compound is selected from the group consisting of 1-deoxynojirimycin, 1-deoxygalactonojirimycin, α -homonojirimycin, 3,4-diepi- α -homonojirimycin, and 4-*epi*-fagomine.

42. (new) The method of claim 41 wherein the compound is 1-deoxygalactonojirimycin.

43. (new) The method of claim 38, wherein the individual has the atypical variant form of Fabry disease.

44. (new) The method of claim 39, wherein the individual has the atypical variant form of Fabry disease.

45. (new) The method of claim 40, wherein the individual has the atypical variant form of Fabry disease.

46. (new) The method of claim 38, wherein the glycosphingolipids are predominantly ceramide trihexoside.

47. (new) The method of claim 39, wherein the glycosphingolipids are predominantly ceramide trihexoside.

48. (new) The method of claim 40, wherein the glycosphingolipids are predominantly ceramide trihexoside.